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Audit to identify the prevalence of urinary incontinence in children and young adolescents attending the Cardiff Paediatric Cystic Fibrosis Unit

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Introduction A number of studies have identified urinary incontinence as a clinical problem in both adult and paediatric cystic fibrosis (CF) patients. Recent studies have reported a prevalence of between 38% and 68% in all adult females with CF and a prevalence of between 4% and 64% in female patients with CF under 25 years of age. Recommendations for the physiotherapy management of CF in the UK state that it should be a clinical standard to identify urinary incontinence in both adults and children with CF and then refer to an appropriate practitioner.
Subjects and Methods An interview-administered questionnaire was used to assess prevalence, severity and impact of urinary incontinence in both male and female paediatric patients over 5 years of age attending the CF clinic at the UHW Cardiff. The questionnaire was administered when patients attended for annual review and 44 (83%) of 53 eligible children and adolescents (aged 5-20.5 years) participated in the audit.
Results 28 males and 16 female CF patients completed the questionnaire. Mean age 13.4 years (range 5.5-20.5 years). 1 male patient (3.6%) and 7 female (43.7%) patients reported urinary incontinence as a current clinical problem. The mean age of patients reporting symptoms was 15.5 years (range 10-18.7 years)
Discussion Patient audit within our clinic population has identified a prevalence of urinary incontinence which is consistent with published data. Results from this audit have informed and improved our annual assessment process and resulted in specialist referral for affected patients. Further audit is required to determine if identification and intervention have resulted in any patients benefit.

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Non invasive ventilation (NIV) in patients with cystic fibrosis (CF) awaiting lung transplantation (LT)

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NIV may be used in patients with CF waiting for LT. The improvement of gas exchange and sleep efficiency due to nocturnal NIV would enable the stabilization of hypercapnic respiratory failure and the improvement of physical performance. Our experience in using NIV in patients with end-stage CF lung disease is reported. 21 patients (mean age 28 ± 10 years), who had carbon dioxide tension in capillary blood ($PcCO_2$) > 50 mmHg and used NIV during the night for at least 15 days were included. A bilevel pressure support ventilator with nasal mask was used and oxygen was administered to obtain a oxygen saturation of haemoglobin between 92 and 95%. The mean value of $PcCO_2$ was 62 ± 11 mmHg at the beginning of NIV. 19/21 patients were on a LT waiting list. The mean duration of NIV support was 220 ± 230 days (range: 18 - 879 days). 14 patients underwent LT (mean waiting time 263 ± 210 days), 5 patients died while awaiting LT and 2 patients were alive. $PcCO_2$ showed a fast and significant increase (> 20 mmHg) in the 5 patients who died. We observed either a gradual increase or a decrease in PcCO2 in 14 patients, who received LT. Nasal bridge ulceration was recorded in 4 patients, who used NIV during both the night and the day. In 3/21 the treatment was not constant because of mask discomfort. In our experience NIV may be administered for a long period and may allow most patients to undergo LT.

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What are the mechanisms that explain exercise capacity in adults with cystic fibrosis

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Aims: The aim was to identify and quantify the independent contribution of each mechanism and then construct an interdependent model to explain exercise intolerance in this population.
Methods: 77 adults with CF underwent comprehensive clinical exercise testing. Exercise capacity, anthropometric measures, CF genotype, *B cepacia* colonization, rest and exercise ventilation, respiratory muscle strength, gas transfer capacity, rest and exercise cardiovascular function, peripheral muscle strength and symptoms were evaluated as independent variables. The 3 largest contributors to exercise capacity, as determined by univariate analysis, were forced into the multiple regression analysis. Multiple linear regression analysis was used to evaluate and quantify the significant contributors to exercise capacity.
Results: Maximal breathing capacity ($R^2 = 0.51$), gas transfer capacity ($R^2 = 0.45$) and lower extremity peripheral muscle strength ($R^2 = 0.48$) consistently demonstrated the greatest independent contribution to exercise capacity ($p<0.001$) and were used to create a model of mechanisms limiting exercise performance. Other statistically significant ($p<0.001$) contributions were made by upper extremity peripheral muscle strength ($R^2 = 0.34$) and maximal exercising heart rate ($R^2 = 0.33$); however, the magnitude of these contributions was noticeably less. No other measures made meaningful contributions to exercise capacity ($p>0.05$). The model of maximal breathing capacity, gas transfer capacity and lower extremity peripheral muscle strength explained 65.5% of the variance seen in exercise capacity.
Conclusions: Exercise capacity is diminished in adults with CF. Ventilation, gas transfer capacity and peripheral muscle strength are the key mechanisms that limit exercise capacity in adults with CF.

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High frequency chest compression (HFCC) therapy waveforms

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Aims: All three waveforms (square, sine and triangle) used for HFCC therapy were developed in Minnesota but as yet no study anywhere has been attempted to compare how these waveforms transmit energy through the chest wall and the lungs.
Methods: We have used Sohn's computer model based on Weibel's airway model (Proc 26th IEEE-EMBS , 2004: 3925-3928) to predict how the waveforms will work on patients. The model used normal lung volumes for a 180 cm male. The computer model airflow at the mouth is a composite of normal breathing and the HFCC compression pulses.
Observations: When the three waveforms were modeled at normal FRC the estimated effectiveness was greatest for the square and least for the triangle waveforms. When the model was changed so each waveform's effect on FRC during operation was entered into the model the estimated effectiveness was greatest for the triangle and least for the sine waveforms. These differences were not affected by the frequencies tested, the limitations of the sine waveform (from 5 to 21 Hertz) were considered.
Conclusions: As yet no one knows which waveform should be prescribed for which patient. Since HFCC was developed for respiratory airway clearance it is vital to study the optimal way to fit the "vest" should fit for HFCC, how to choose the waveform to prescribe, the frequencies to use, the duration of use for each frequency, the times per day for therapy and how long each therapy session should last. Some of these questions can be "solved" by computer modeling; the results of these predictions will have to be tested on patients. We will investigate the three waveforms, how each operates in our future researches to predict what human studies may find such as similarities and differences between model and patients.